A CLINICAL STUDY OF PAPILLOEDEMA

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CERTIFICATE

This is to certify that this dissertation entitled **A study on etiology of papilloedema in South Tamilnadu** submitted by **Dr. M.Rita Hepsi Rani** to the faculty of Ophthalmology The Tamil Nadu Dr. MGR Medical University, Chennai in partial fulfillment of the requirement for the award of M.S Degree Branch III (Ophthalmology), is a bonafide research work carried out by her under our direct supervision and guidance.

Dr.R .Mani.,

Additional Professor of Ophthalmology,
Tirunelveli Medical College,
Tirunelveli.

Dr.A .Meenakshisundaram.,

Prof & Head of the Department
Department of Ophthalmology
Tirunelveli Medical College,
Tirunelveli.

THE DEAN
Tirunelveli Medical College,
Tirunelveli.

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PAPILLOEDEMA

INTRODUCTION

The etiological diagnosis and management of papilloedema is one of the most fascinating and perplexing problems of Neuro ophthalmology. Papilloedema is one of the most common conditions an ophthalmologist comes across in routine clinical practice. The ophthalmologist has a vital role in differentiating papilloedema from pseudo papilloedema, in confirming the diagnosis and in advising the time of surgery and in giving the prognosis as far as vision is concerned. In this study an attempt has been made to study papilloedema in all its aspects.

AIM OF THE STUDY

1. To study the different etiologies of papilloedema in a tertiary level hospital.

2. To study the progression of papilloedema during the course of disease.

3. To find out the associated systemic causes.

REVIEW OF LITERATURE

Parson in 1908, first applied the term "papilloedema" to swellings of the optic disc that exceeded two diopters and were associated with increased intra cranial pressure.

The German expression "Struungspapille" is used in this context. Lesser degrees of disc swelling with increased intra cranial pressure were designated as Papilloderm (Uthoff). Hubler recommends that the term Struungspapille be extended to encompass all stages of disc swelling caused by increased intra cranial pressure.

In 1911, Paton and Holmes made the first detailed morphologic studies of disc swelling and provided definite description of non inflammatory "passive oedema" encountered in patients with normal vision and increased intra cranial pressure. For these cases, they used Parson's term "Papilloedema" and the term "Optic neuritis" to swelling of the disc associated with local inflammation and loss of vision.

The term "Papilloedema" is used for all forms of disc swelling that result from increased intra cranial pressure. And the unrestricted expression "oedema" of the optic disc or "disc

oedema" to all other forms of disc swelling of local or systemic etiology that do not significantly affect vision.

It is nearly always bilateral, although it may be asymmetrical all patients with papilloedema should be suspected of having an intra cranial mass unless proved otherwise. However not all patients with raised intra cranial pressure will necessarily develop papilloedema. Tumours of cerebral hemispheres tend to produce papilloedema later than those in the posterior fossa. Patient with a history of previous papilloedema may develop a substantial increase in intra cranial pressure but fail to re-develop papilloedema because of glial scarring of the optic nerve head.

DEFINITION

Papilloedema is defined "as bilateral passive non inflammatory edema of the optic disc due to raised intracranial pressure which is almost always bilateral and without visual deficit".

Normal CSF pressure in the lateral recumbent position varies between 100 – 250 mm of H_2O. Normal opening pressure of CSF on lumbar puncture is < 80mm H_2O in infants, < 90mm H_2O in children and < 210mm H_2O in adults.

ANATOMY OF OPTIC NERVE HEAD

(a) REGIONS:

The optic nerve head consists of four regions

i) Superficial nerve fibre layer

ii) Prelaminar region

iii) Laminar region

iv) Retro laminar region.

i) Superficial nerve fibre layer

This part is essentially composed of axonal bundles of nerve fibres of the retina (94%) which converge on the optic disc and astrocytes (5%). The optic disc is covered by a thin layer of astrocytes, the internal limiting membrane of Elsching which separates it from the vitreous and is continuous with the internal limiting membrane of retina. When the central portion of this membrane is thickened it is referred to as the central meniscus of Kuhnt[2].

All the layers of retina, apart from the nerve fibre layer (NFL) near the optic nerve, are separated from it by a partial rim of glial tissue called the intermediate tissue of Kuhnt.

ii) Prelaminar region:

The predominant structures at this level are neurons and a significantly increased quantity of astroglial tissue. The border tissue of Jacoby separates the nerve from choroid.

iii) Laminar region:

It is a fibrillar sieve like structure made up of fenestrated sheets of scleral connective tissue lined by glial tissue. The bundles of optic nerve fibres leave the eye through these fenestrations.

iv) Retro laminar region:

This area is characterized by a decrease in astrocytes and the acquisition of myelin that is supplied by oligodentrocytes. The addition of myelin sheath nearly doubles the diameter of optic nerve from 1.5mm to 3.0mm as it passes through sclera.

(b) BLOOD SUPPLY:

(i) Surface nerve fibre layer is mainly supplied by the capillaries derived from the retinal arterioles, which anastamose with vessels of the prelaminar region. Occasionally a ciliary derived vessel from the pre laminar region may enlarge to form the cilioretinal artery[2].

(ii) The prelaminar region is supplied by vessels of ciliary region. These vessels are primarily derived from the peripapillary choroidal system or from separate branches of the short posterior ciliary arteries.

(iii) The lamina cribrosa region is supplied by the ciliary vessels which are derived from the short posterior ciliary arteries and arterial circle of Zinn-Haller.

(iv) The retro laminar region is supplied by both the ciliary and retinal circulation with the former coming from recurrent pial vessels. The central retinal artery provides centripetal branches from the pial plexus and also centrifugal branches.

The disc and retina are exposed to intra ocular pressure whereas the retro laminar and proximal nerve to cerebrospinal fluid pressure (Hayreh and Dass 1960, Hayreh 1963 & 1974).

The optic nerve is surrounded by meningeal sheaths dura, arachnoid and pia matter upto lamina cribrosa. There is an extension of the intra cranial subarachnoid space forward around the optic nerve to the back of the eye ball. So any increase in ICP compresses the thin walled retinal veins, as they cross the extension of subarachnoid space leading to congestion and bulging of the disc. Since subarachnoid space around optic nerve is continuous with the intracranial subarachnoid space, both eyes will exhibit papilloedema. Optic disc does not possess the cells of Muller, which holds the nerve fibres together, hence it easily swells in papilloedema.

(c) AXOPLASMIC FLOW:

Normally there is a continuous flow of axoplasm along the optic nerve.

1. Antegrade flow or orthograde transport:

This is the flow from the retinal ganglion cells

along the optic nerve axonal fibres to their terminals in the lateral geniculate body[1].

- **Slow component: proteins and enzymes**

The slow component materials move at a speed of 1-3 mm per day. It is driven through peristaltic waves that passively drive across the content and it is the component that is interfered early in the onset of increased intra cranial pressure.

- **Rapid component**: (Subcellular organelles, mitochondria)

Materials move at a speed of 400 – 1000mm/day.

The velocity of the axoplasmic flow in the fast component depends upon the intra axonal pressure which is determined by the difference between the IOP gradient in the pre-laminar area and optic nerve tissue pressure gradient in the post laminar area. The optic nerve tissue pressure in turn is determined by CSF pressure in the dural sheath[3].

2. Retro grade flow:

Lysosomes and mitochondria flow from the lateral geniculate body to the retinal cells also occurs at an intermediate rate.

Mucopolysaccharides, proteins, glycolipids, gangliosides, phospholipids, cholesterols, smooth endoplasmic reticulum and associated elements constitute axoplasmic material. Hayreh injected labelled aminoacid like leucine or protein into the

vitreous cavity of monkey and demonstrated accumulation in both fast and slow components in the lamina cribrosa with increase in ICP. The accumulation of axoplasm results in swelling of axons as seen by electron microscopy.

Radius and Anderson (1980) demonstrated impaired slow axonal transport and axonal swelling by injecting the drug β-β' imino di propionitrite (IDPN) that selectively and directly blocks axonal transport producing optic disc swelling in dogs, guinea pigs and in monkeys.

1. Papilloedema occurs only when there is patency of meningeal space surrounding the optic nerve and intra cranial structures. Blockage of this space by adhesion, or tumours prevents papilloedema from occurring on the side of obstruction.
2. Papilloedema does not occur in eyes in which antecedent optic atrophy had destroyed most or all of the nerve fibres.
3. Most importantly slow axonal transport is clearly abnormal in patient with papilloedema as well as in patient with disc swelling from all other causes (Ischemia, inflammation, hypotony etc).

PATHOGENESIS

a) Intra ocular pressure and optic nerve tissue pressure:

i) Intra axonal pressure - which is determined by the difference between the intra ocular pressure gradient in the prelaminar area and optic nerve tissue pressure gradient in the post laminar area.

ii) The optic nerve tissue pressure:

Which in turn is determined by the CSF pressure in the dural sheaths.

Intra axonal pressure = IOP in prelaminar area - optic nerve tissue pressure in post laminar region.

Normally the IOP (14-20mm Hg) is higher than the tissue pressure in the optic nerve (6 – 8mm Hg). This pressure differential is the force driving the axoplasm in the region of the lamina cribrosa. Hence a fall in the IOP or an increase in the optic nerve tissue pressure following a rise in the CSF pressure will interfere with the axoplasmic flow, leading to stasis and accummulation of the axoplasm.

Experimentally this was proved in a successful animal model developed by Hayreh. He produced papilloedema in

monkeys by slowly inflating a balloon that had been placed in the sub arachnoid space within the skull, thus simulating a space occupying lesion. Later he demonstrated the regression of the papilloedema on the same animal model by opening the dura and arachnoid around the optic nerve on one side. This forms the basis of the surgery for relieving the papilloedema viz, optic nerve sheath fenestration.

In another study Hayreh injected fluorescein solution in to the CSF of the monkey and observed fluorescence of the optic disc.

This accumulation of axoplasm results in the swelling of axons seen by electron microscopy. Accumulation is mainly due to blockage of slow axonal transport.

The cranial cavity is an almost rigid bony enclosure completely filled by tissue, CSF and circulating blood. Within this enclosure CSF is constantly produced at the rate of 0.37ml per minute primarily inside the ventricular system.

b) CIRCULATION OF CSF

CSF is secreted by choroid plexus of lateral, third and fourth ventricles and ependymal lining of the ventricular system. CSF flows from the lateral ventricles through the inter ventricular foramen into the third ventricle and then to the fourth ventricle through the cerebral aqueduct of Sylvian and

out into the sub arachnoid space through the foramina of Luschka and Magendie. In the subarachnoid space, CSF flows rostrally from the posterior fossa through the lower ventral basal cisterns and tentorial notch to reach the inter peduncular and chiasmatic cisterns. The CSF then flows dorsally through the communicating cisterns to reach the dorsal cisterns, and lateral and superiorly from the chiasmatic cisterns into the cistern of the Sylvian fissure. From the cisterns and Sylvian fissures, CSF moves outward and superiorly over the cerebral convexities where it is absorbed.

The chief route of absorption of CSF is through the arachnoid granulations that protrude into the venous sinuses and the diploic veins. These vessels drain into the internal jugular vein and extra cranial veins.

CHART – 1

Choroid plexus

↓

Lateral ventricles

↓

Inter ventricular foramen

↓

3rd ventricle

↓

Cerebral aqueduct of Sylvius

↓

4th ventricle

↓

Foramen of Luschka & Magendie

↓

Subarachnoid space

↓

Arachnoid granulations of

Venous sinus /diploic vein

Under appropriate circumstances, as little as 80cc of rapidly added volume (CSF, blood, oedema, tissue) will raise ICP to a level incompatible with life.

CLINICAL FEATURES OF PAPILLOEDEMA

SYMPTOMS AND SIGNS

Table 1

Non visual manifestations	**Visual manifestations**
1. Head ache – Early symptoms. Due to stretching of meninges	1. Transient visual obscuration.
2. Nausea and projectile vomiting – due to increase in ICP.	2. Loss of central vision
3. Bradycardia and respiratory failure due to herniation of medulla in foramen magnum.	3. Diplopia
4. Loss of consciousness due to compression of cerebral cortex and decrease in blood flow.	4. Visual field defects
5. Generalised motor rigidity – due to crowding of temporal lobe, as a result of herniation of the hippocampal gyrus causes pressure on the crus cerebri.	
6. Dilated pupil not responding to light – due to direct pressure on the oculomotor nerve or dorsal midbrain.	

VISUAL MANIFESTATION:

1. Transient visual obscuration:

Transient obscuration of vision may occur in both eyes simultaneously with rapid recovery usually lasting seconds but sometimes lasting hours.

Patients may experience upto 20-30 attacks per day, with obscurations precipitated by change of posture, particularly from sitting to the standing position or from the lying down to sitting or standing position.

i. The cause of these transient obscuration is related to transient compression or ischaemia of the intra cranial portion of the optic nerve or optic chiasm by a distended third ventricle.

ii. Ischaemia of optic nerve head and downward herniation of the hippocampal gyrus in the tentorial notch.

2. Visual field defects:

(i). Concentric enlargement of the blind spot is the most common and frequently the only visual field defect observed is patient with papilloedema.

The cause of the enlarged blind spot is compression, detachment and lateral displacement of the peripapillary retina. The enlarged blind spot represents a refractive scotoma caused by hyperopia of the peripapillary retina.

Other visual field defects are

(ii) concentric constriction

(iii) homonymous hemianopia

iv) central and arcuate scotoma

v) complete blindness. Inferior nasal quadrant was most frequently affected than temporal quadrant. Thus an eye may be left with only a temporal island of vision before progressing on to complete blindness. (vi) constriction of the visual field is late sign which occurs in chronic papilloedema as it progresses to optic atrophy.

3) Loss of central vision

Acute or progressive loss of central vision may be due to

i) Haemorrhage or exudates at the macula.

ii) Increased optic nerve tissue pressure causing occlusion of prelaminar anterioles.

iii) The underlying etiological condition causing retinal vascular occlusion.

4) Diplopia

This may result from compression or stretching of the abducent nerve at the base of the skull which may be unilateral or bilateral (false localizing sign).

Very occasionally trochlear nerve palsy may also occur, presumably from compression of the dorsal midbrain or direct

compression of the nerves themselves by a ballooned supra pineal recess.

OPHTHALMOSCOPIC APPEARANCE:

As suggested by Hughlings and Jackson (1871) and by Bolloyt and Beecton (1966) papilloedema may be classified into four types (a) early (b) fully developed (c) chronic (d) atrophic.

a) Early papilloedema:

The early phase of papilloedema consists of the incipient disc changes that occur before the development of obvious disc swelling. The features include, hyperemia of the optic disc blurring of the peripapillary retinal nerve fibre layer, swelling of the optic disc, blurring of the disc margins, peripapillary flame shaped haemorrhages and absent spontaneous venous pulsations.

b) Fully developed papilloedema:

The characteristics are, increase in disc swelling (2-6 diopters), venous engorgement. Numerous splinter haemorrhages, microaneurysms, partial obscuration of traversing blood vessels, obliteration of physiological cup, vessels obscured by nerve fibre layer edema, peripapillary flame shaped haemorrhages, cotton wool spot which are due to focal retinal infarcts, tortuous vessels surrounding the disc.

Severe cases may show circumferential retinal folds, Patons lines (peripapillary chorio–retinal folds), hard exudates and haemorrhage in macula giving macular fan appearance.

c) Chronic papilloedema:

This is characterized by:

Resolved haemorrhages, exudates and edema over the disc along with loss of the central cup. The disc develops a rounded appearance. There is appearance of pallor of the disc. Optic discs are markedly elevated with a champagne cork appearance. Opto-ciliary shunts and drusen may occur. Most patients with chronic papilloedema have evidence of nerve fibre layer atrophy. The atrophy ranges from slit like defects to diffuse loss which is readily appreciated by red free filter of direct ophthalmoscopy or slit lamp bio microscopy.

d) Post papilloedemic optic atrophy (secondary optic atrophy)

Characteristics are:

Grey white pallor of disc with blurred margins.

Attenuated sheathed vessels.

Gliosis on the disc surface and margin.

Frisen grading system is most useful as it classifies the papilloedema grade by severity.

FRISEN GRADING SYSTEM

Table - 2

Stage 0:Normal optic disc Blurring of nasal, superior, and inferior poles in inverse proportion to disc diameter. Radial nerve fibre layer (NFL) without NFL tortuosity. Rare obscuration of a major blood vessel, usually on the upper pole.
Stage 1:Very early papilloedema Obscuration of the nasal border of disc. No elevation of the disc borders. Disruption of normal radial NFL arrangement with greyish opacity along NFL bundles Normal temporal disc margin. Subtle greyish halo with temporal gap (best seen with IDO) Concentric or radial retro choroidal folds.
Stage 2:Early papilloedema Obscuration of all borders Elevation of the nasal border Complete peripapillary Halo
Stage 3:Moderate papilloedema Obscuration of all borders Increased diameter of optic nerve head Obscuration of one or more segments of major blood vessels leaving the disc peripapillary halo irregular outer fringe with finger like extensions
Stage 4:Marked papilloedema Elevation of the entire nerve head Obscuration of all borders Peripapillary halo Total obscuration on the disc of a segment of a major blood vessel.
Stage 5:Severe papilloedema Dome shaped protrusion representing anterior expansion of the optic nerve head. Peripapillary halo is narrow and smoothly demarcated. Total obscuration of a segment of a major blood vessel may or may not be present. Obliteration of the optic cup.

ETIOLOGY

Mechanism of increased ICP

1. An increase in the intra cranial tissue by a space occupying lesion.
2. An increase in intra cranial tissue volume by focal or diffuse cerebral edema.
3. A decrease in total volume within the cranial vault by thickening of the skull.
4. Blockage of the flow of CSF within the ventricular system or within the arachnoid granulations.
5. Reduced absorption of CSF from obstruction or compromise of venous flow both intra cranially and extra cranially.
6. Increased production of CSF by an intra cranial tumour at a rate that precludes absorption for maintenance of normal ICP.
    - Intra cranial masses
    - Aqueductal stenosis
    - Brain abscess
    - AV malformation
    - Trauma

- Acute and chronic subdural haematoma
- Epidural hematoma
- Subarachnoid haemorrhage
- Septic and aseptic meningitis and encephalitis
- Carcinomatous / lymphomatous / leukemic meningitis
- Cerebral and leptomeningeal gliomatosis
- Spinal cord lesions
- Landry Guillain – Barre syndrome
- Chronic inflammatory demyelinating polyneuropathy (CIDP)
- Mucopolysaccharoidosis
- Cranio synostoses
- POEMS syndrome
- Multiple myeloma
- Hyperthyroidism
- Late onset citrullinemia
- Pseudo Tumor Cerebri

1. Intra cranial masses

Intra cranial masses produce raised ICT by several mechanisms

i) They may act solely as space occupying lesions

ii) They may produce focal or diffuse cerebral edema

iii) They may block the CSF drainage pathways by direct compression or infiltration of the arachnoid villi or cerebral venous sinuses.

iv) Producing increased protein and blood products that secondarily block the arachnoid villi.

Papilloedema does not develop in every patient with a cerebral tumor.

Infra tentorial tumours are more likely to produce papilloedema than supra tentorial tumours.

An intracranial tumour in any position may induce papilloedema , the highest percentage being found with tumours of the midbrain, parieto occipital region and cerebellum. Papilloedema due to tumours of the anterior fossa is relatively rare and occurs late in the course of the disease. In general, those tumours which tend to produce internal hydrocephalus are most likely to cause papilloedema. The site of the tumour is more important than its nature, size or rate of growth.

Papilloedema may occur with any type of intracranial mass including primary and secondary metastatic brain tumours. Hamartomas, teratomas, giant aneurysms, A.V. malformations, cysticercus cysts, and granulomas (eg: in syphilis, sarcoidosis or tuberculosis).

According to Stopford (1928) supra tentorial tumour rarely cause primary obstruction to the flow of CSF through the aqueduct of Sylvius, instead such tumors produce papilloedema by deflection of the falx and pressure upon the great vein of Galen.

Tumors below the tentorium produce papilloedema by obstruction of aqueduct. Some infra tentorial masses do not obstruct the aqueduct, but still produce papilloedema. Stopford (1928) suggested that elevation of tentorium with pressure on the vein of Galen is responsible.

2. Obstruction to flow of CSF:

Aqueductal stenosis: This often presents in childhood.

It may be congenital which may or may not be associated with Chiari malformation or acquired from intracranial infections such as toxoplasmosis or mumps ependymitis. Sometimes it can present in infants with microcephaly, in adult with head ache, dorsal mid brain syndrome, meningitis, or endocrine disturbances from the compression of the pituitary gland, seizures, gait disturbances and CSF rhinorrhea. Neuro imaging shows marked enlargement of lateral and third ventricles with normal fourth ventricle. The symptoms are relieved by ventriculo peritoneal shunt.

3. Trauma:

i) Papilloedema occurs in 20 – 30% of persons who have suffered severe head trauma. The increased ICP is caused by a severe subarachnoid haemorrhage or a significant intra cerebral subdural or epidural haematoma. It may also be caused in some cases by cerebral venous sinus thrombosis or diffuse or localized cerebral oedema. Papilloedema may occur immediately or several days after injury or up to 2 weeks later.

ii) Subarachnoid haemorrhage:

This usually produces papilloedema by blocking CSF flow within the ventricular system or blocking CSF absorption at the arachnoid granulations.

iii) Papilloedema from acute and chronic subdural hematoma. Papilloedema occurs in patient with both acute and chronic subdural hematoma. Gardner proved that papilloedema occurs early in children but late in adult, similarly most often occur in acute subdural hematoma than chronic subdural hematoma.

iv) Epidural hematoma:

Stevenson et al (1964) reported the case of a patient who developed papilloedema from an epidural hematoma that occurred several weeks after a vertex skull fracture.

Papilloedema resulted from compression of the superior sagittal sinus in this case.

4. Septic and aseptic meningitis and encephalitis:

Meningitis produces papilloedema by (a) diffuse cerebral edema, (b) obstruction to CSF flow by aqueductal stenosis or (c) obstruction of CSF resorption through arachnoid granulations. Papilloedema occurs in 2.5% of patients with meningitis, more so in tubercular meningitis (25% of cases).

Approximately 19% of patients with viral encephalitis have papilloedema. Herpes simplex, Herpes zoster, and measles encephalitis are commonly associated with papilloedema whereas rubella, mumps and varicella are not associated with papilloedema. Patient with poliomyelitis may develop increased intracranial pressure during both acute and convalescent phases of the disease.

Infection:

In almost all CNS infection and inflammation, swelling of the optic disc may occur without elevated ICP, presumably from inflammation of the optic nerve.

5. Elevated venous pressure:

Obstruction of cerebral venous drainage may result in increased ICP and papilloedema. The obstruction is most often caused by compression or thrombosis with the vessels more

commonly being affected are the superior sagittal and transverse (lateral) sinuses. The common causes are:

Tumours – those obstructing the superior sagittal sinus are usually extra axial like meningiomas. The transverse sinuses can be occluded by acoustic neuromas, meningiomas and metastatic tumours[1].

Septic thrombosis of the transverse sinus occurs in the setting of acute or chronic otitis media in which there is extension of the infection to the mastoid air cells and then to the adjacent transverse sinus.

Septic thrombosis of the cavernous sinus also can produce papilloedema but late in the course of the process.

6. Extra cranial lesions:

i) Tumours of the spinal cord may produce increased ICP by two mechanisms.

a) Some tumours may be located in the high cervical region and the upward swelling of the tumour with compression of the cerebellum and obstruction of the CSF flow through the foramen magnum.

b) The majority of the tumours of the spinal cord associated with papilloedema are however ependymomas and neurofibromas located in the thoracic and lumbar regions. These tumours produce extremely high concentrations of protein in the CSF and

the papilloedema is probably a result of obstruction of the drainage of CSF at the arachnoid granulations by the proteins.

ii) Landry Guillain - Barre syndrome (GBS)

Proteins in the arachnoid villi and granulations alter the cerebral venous dynamics or cause partial venous thrombosis leading to increased ICP.

iii)Chronic inflammatory demyelinating polyneuropathy (CIDP) causes increased ICP by increasing in the protein concentration which is the hallmark of the disease.

iv) The POEMS syndrome is an unusual multi system disorder characterized by polyneuropathy (P), organomegaly (O) endocrinopathy (E), monoclonal gammopathy (M) and skin changes (S). Patients with POEMS syndrome occasionally develop increased intra cranial pressure associated with papilloedema.

7. Post convulsive papilloedema

Low grade or transient papilloedema has been observed after a long series of convulsions (status epilepticus).

8. Edema of optic disc with systemic diseases

i) Cardiopulmonary insufficiency, chronic respiratory acidosis, hypercapnia, and polycythemia cause an increase in

the venous pressure which leads to increased intracranial pressure and thereby papilloedema.

ii) Pulmonary emphysema leads to respiratory acidosis which causes an accumulation of CO_2 in brain tissue reflected by inversion of the normal CO_2 tension ratio between cerebrospinal fluid and arterial blood. This in turn causes dilatation of cerebral capillaries and increases intra cranial blood volume.

iii) Extensive obesity as in Pickwickian syndrome causes ↓ vital capacity, polycythemia, cyanosis along with bilateral disc edema.

iv) Hypoparathyroidism and papilloedema

Both idiopathic and acquired forms of hypoparathyroidism are associated with papilloedema of mild degree.

v) Thrombocytopenic purpura and edema of optic disc.

vi) Leukemia and papilloedema

9. Arterio Venous Malformation:

AVMs need not rupture to produce papilloedema. The mechanism producing increased ICP is by direct shunting of arterial blood from AVMs, into cerebral venous system leading to increased cerebral venous pressure and decreasing CSF

absorption. Both intra cerebral and dural based AVMs involving the transverse sinus are associated with papilloedema.

10. Others:

Non infective inflammatory

i) Sarcoidosis - Papilloedema may be caused by aseptic meningitis or dural venous sinus thrombosis.

ii) Behcet's - Pseudo tumour cerebri with papilloedema is the most common presentation of dural venous sinus thrombosis in Behcet.

11. Inborn errors of metabolism

- Mucopolysaccharoidosis – obstruction of CSF over cerebral convexities, or in the arachnoid villi or both by deposits of mucopolysaccharide in the meninges, leading to delayed CSF absorption.
- Citrullinemia – is caused by deficient activity of argino succinate synthetase, a urea –cycle enzyme. This disorder may present acutely in neonates, sub acutely in children or as a late onset condition in young adults. When the condition is of the late – onset type, it may produce increased intra cranial pressure associated with papilloedema.
- Craniosynostosis
- Plasma cell dyscrasia

- Toxins – (eg) lead can damage Blood brain barrier, cause severe cerebral edema and ↑ICP
- Hyperthyroidism – due to compression or ischaemia of optic nerve caused by dysthyroid orbitopathy
- Pseudo Tumor Cerebri

12. Pseudo Tumour Cerebri (Benign Intra cranial hypertension / Idiopathic intra cranial Hypertension)

This syndrome was first recognized by Quincke (1897). Warrington (1914) was the first to use the term "Pseudo tumour cerebri". Foley introduced the term "Benign intra cranial hypertension".

Clinical manifestation:

The most common presenting symptom in patients with pseudo tumour cerebri is head ache, occurring in more than 90% of cases. Headache is usually generalized, worse in the morning and aggravated when cerebral venous pressure is increased by some type of Valsalva manoeuvre (coughing, sneezing etc)

- nausea, vomiting, dizziness, and pulsatile tinnitus
- visual manifestations like

(a) transient visual obscuration

(b) loss of vision from macular haemorrhages, exudates, pigment epithelial changes, retinal striate, choroidal

folds, sub retinal neovascularization or optic atrophy

(c) horizontal diplopia from unilateral or bilateral abducent nerve palsy and rarely

(d) Vertical or oblique diplopia from trochlear nerve paresis, oculomotor nerve paresis or skew deviation.

Aetiology of Pseudo Tumor Cerebri

Usually idiopathic in over 90% of cases. Pseudo tumour cerebri occurs in young obese women and occasionally men with no evidence of underlying disease.

In about 10% patients, particularly in men and non obese young women, Idiopathic Intra cranial Hypertension develops in a number of clinical settings namely.

Obstruction and impairment of cerebral venous drainage. Endocrine and metabolic dysfunction viz, Addisons disease, hypoparathyroidism, hyperthyroidism, hypothyroidism, menarche, menopause, obesity (idiopathic) pregnancy, exposure to drugs like corticosteroids, oral contraceptives, tetracycline, nalidixic acid, amiadrone, VitA, systemic illness like anaemia, chronic respiratory insufficiency / hypertension, multiple sclerosis, Systemic Lupus Erythematous, and withdrawal of certain drugs (eg) steroids.

PATHOLOGY

The pathology of papilloedema shows signs of passive edema without evidence of inflammation. The oedematous changes are located in the optic nerve head in front of lamina cribrosa. The small blood vessels are engorged and tortuous. The nerve fibres in the optic nerve head are swollen and axoplasmic stasis is noted in them. The physiological cup gets filled in and the internal limiting membrane is raised. The nerve fibres become swollen and varicose and ultimately degenerate. They shows numerous cytoid bodies in front of lamina cribrosa. Electron microscopy shows engorgement of axons in the laminar portion of the optic nerve. The swollen axons are filled with mitochondria. The neuroglia proliferates and the mesoblastic tissue around the vessels becomes thickened. Retinal exudates distributed radially along the folds may be present in the macular region corresponding to the clinical appearance of a macular fan or macular star. The separation of the swollen optic disc from adjacent retina is in the form of an 'S' shaped curve. Compression, detachment and lateral displacement of the peripapillary retina appears to be reason for enlarged blind spot[4].

DIAGNOSIS

The most important method of diagnosing papilloedema is careful ophthalmoscopic examination. 1) red free ophthalmoscopy and 2) slit lamp bio-microscopy with a hand held lens or contact lens. If the diagnosis remains uncertain several options are available. 3) Fluorescein angiography is often used to confirm early papilloedema. 5ml of 10% fluorescein sodium is rapidly injected into a superficial arm vein. The passage through retinal vessels is then observed and photographed at rapid intervals with a cobalt blue filter interposed in front of the light source to visualize the fluorescence of the dye. The early phase shows capillary dilatation dye leakage and micro aneurysm formation. Later films show leakage of dye beyond the disc margins. 4) Orbital echography is useful in cases of questionable papilloedema. This sensitive test determines of the diameter of the optic nerve. 5) By performing A and B scan ultrasonography we can determine optic disc edema and optic disc drusen. 6) CT scan and MRI can be used to detect evidence of an intra cranial mass or hydrocephalus.

High resolution MRI of the orbit shows characteristic changes of the optic nerves and posterior globe in papilloedema. The Subarachnoid space becomes distended, the nerve sheath widens and there is flattening of the posterior sclera. The prelaminar optic nerve may enhance and protrude anteriorly.

COURSE

The rapidly developing papilloedema depends to a large extent on the etiology and degree of the increased ICP.

Clinically detectable papilloedema is not usually present with acute elevation of ICP, and may take over a week to develop. Cases have been reported of papilloedema within 2 - 4hours after intra cranial haemorrhage.

Fulminant haemorrhagic papilloedema was also seen within 5 – 8 hours in conditions like metastatic frontal lobe tumour and epidural haematoma.

In addition, minimal papilloedema may exist and suddenly become fully developed in several hours in certain settings such as encephalitis with development of cerebral abscess.

Fully developed papilloedema may resolve completely within hours, days or weeks depending on the way in which ICP is lowered.

In humans, papilloedema can resolve 6 – 8 weeks after a successful craniotomy to remove a brain tumour. Optic disc edema may improve in less than a week following optic nerve sheath decompression.

In most cases the retinal venous as well as disc capillary dilatations begin to regress as soon as ICP is lowered to a normal level. During the next few days to weeks disc hyperemia and elevation resolve. The last abnormalities to disappear are blurring of the disc margins and abnormalities of the peripapillary retinal nerve fibre layer.

In severe cases papilloedema resolves into optic atrophy. Optic atrophy following papilloedema is referred to as 'Secondary' optic atrophy.

DIFFERENTIAL DIAGNOSIS

Pseudo papilloedema:

This reflects differing morphological appearance of the disc.

1. Anamolously elevated optic disc.

i) Small hypermetropic disc, tilted optic disc, hypoplastic optic disc

ii) Myopic disc that are asymmetrical

iii) Optic nerve head drusen especially burried drusen which may be mistaken for early papilloedema. However unlike true papilloedema, there is no obscuration of superficial fine vessels at the edge of the disc. Auto fluorescence may be positive which helps in the diagnosis.

2. Medullated nerve fibres around the disc.

3. Bergmiester's papilla

4. Bilateral disc swelling may be caused by any one of the following

- malignant hypertension
- bilateral papillitis
- bilateral compressive thyroid ophthalmopathy

- bilateral simultaneous anterior ischaemic optic neuropathy
- bilaterally compromised venous drainage in CRVO or carotico cavernous fistula.

5. Optic perineuritis

6. Infiltrative optic neuropathy eg: from leukemia

7. Compressive optic neuropathy eg: from optic nerve sheath meningioma.

TREATMENT

The main stay of treatment is to reduce the increased Intra cranial Pressure. If timely decompression is successfully performed, the effect is remarkable. Along with relief of the general symptoms like head ache, vomiting, stupor etc, vision improves rapidly (unless the nerves have been irreversibly damaged) and papilloedema subsides.

ICP may be reduced by medical / surgical management

(i) If increased ICP is directly related to a mass lesion – Removal of the lesion is the treatment of choice.

(ii) If the lesion cannot be removed or if CSF absorption is reduced at the level of arachnoid villi – Shunting of CSF into atrial or peritoneal cavities

(iii) If there is cerebral edema, osmotic agents diuretics or corticosteroids may reduce swelling particularly in acute period.

(iv) If papilloedema is due to pseudo tumour cerebri, it is medically managed

(v) Surgical treatment

Treatment of Pseudo Tumor Cerebri includes:

1. Withdrawal of causative drugs.
2. Weight reduction.

3. Medical therapy

T. Acetazolamide 1gm/day – divided doses 250mg Qid or 500 mg B.D

This reduces production of CSF by inhibition of carbonic anhydrase

↓

↓ Na ion transport across the choroidal epithelia.

↓

↓ CSF production

Side effect:

Parasthesia, numbness, lethargy

decreased libido, metallic / dry taste in mouth.

4. Repeated lumbar punctures - are advocated as non medical, non surgical method of relieving the increased ICP of Idiopathic Intra Cranial Hypertension. The theory behind is that the needle used for lumbar puncture creates an opening in the dura through which CSF leaks. With several lumbar puncture, one creates a sieve that allows sufficient egress of CSF that ICP is normalized. This procedure is painful, technically difficult to perform and causes a low pressure headache.

Surgery is considered under the following circumstances.

1. Progressive loss of vision despite maximal medical therapy.

2. Severe or rapid vision loss at onset, including the development of an afferent pupillary defect or signs of advanced optic nerve dysfunction.
3. Severe papilloedema causing macular edema or exudates. Surgical procedure includes (a) optic nerve sheath decompression (ONSD) and (b) CSF diversion procedures.

Hayreh (1969) initiated optic nerve sheath decompression through lateral orbitotomy.

a) Optic nerve sheath decompression: ONSD

In this procedure, a window or multiple slits are made in the dural sheath of the optic nerve immediately behind the globe. A successful ONSD results in resolution of papilloedema on that side and occasionally on the other side with improvement in the visual function in many cases. The procedure immediately reduces pressure on the nerve by creating a filtration apparatus that controls the intra vaginal pressure surrounding the orbital segment of the optic nerve.

Complication of ONSD are:

1. Extra ocular motility dysfunction – usually transient.
   Usually involves the lateral rectus muscle.
2. Pupillary dysfunction
3. Loss of vision from vascular occlusion
   a) CRAO – Central Retinal Artery Occlusion

b) BRAO – Branch Retinal Artery Occlusion

c) Choroidal ischaemia / infarcts -

4. New visual field defect
5. Orbital haemorrhage
6. Transient or protracted blindness
7. Globe perforation

b) CSF diversion procedures

A CSF diversion procedure treats Idiopathic intra cranial Hypertension by lowering the intra cranial pressure.

Lumbo peritoneal shunt is more commonly performed than ventriculo peritoneal shunting.

LP shunt in which a silicone tube is placed percutaneously between the lumbar sub arachnoid space and the peritoneal cavity.

Complication of shunting includes:

1. Shunt valve or tubing obstruction
2. Over shunting (low pressure head ache)
3. Catheter migration / abdominal pain
4. Infection
5. CSF leak
6. Cerebellar tonsillar herniation (acquired Chiari malformation)

MATERIALS AND METHODS

The study of etiologies and ophthalmoscopic findings, CT findings of papilloedema in South Tamil Nadu was carried out in the Department of Ophthalmology, Tirunelveli Medical College Hospital, Tirunelveli.

Settings:	Ophthalmology ward, Neuro Medicine ward, Neuro surgical ward, Medical ward, Pediatric ward.
Study design:	Single centre observational prospective hospital based study.
Period of study:	June 2007 to June 2009.
Ethical approval:	Obtained

Tirunelveli Medical College Hospital is a tertiary care centre in South Tamil Nadu. The patient population is fairly representative sample of the disease pattern in this region.

75 patients with papilloedema in the above period who satisfied the set criteria were included.

1). Papilloedema is defined as bilateral passive non-inflammatory edema of the optic disc due to raised intra cranial pressure which is almost always bilateral and without any visual deficit.

2). The patients presented with headache, nausea, vomiting, defective vision with ophthalmoscopic findings of fundus with papilloedema.

3). 75 patients, with visual acuity recording, field charting, colour vision, neurological examination, CT brain were included.

Exclusion criteria:

1). Inflammatory optic disc edema.

2). Unilateral papilloedema were excluded.

3). Pseudo papilloedema were excluded.

After obtaining consent from either patient or relatives, all patient in the study group were evaluated by complete medical history, full ophthalmoscopic examination, fundus examination and CT scan and MR imaging of brain in indicated cases.

Clinical history:

Clinical history was recorded from either the patient or his/her relatives. Special emphasis was given to the presenting complaint, mode of onset, duration, associated features, activity of the patient at the time of onset and the pattern of presentation.

Presence or absence of risk factors for papilloedema were also noted. Past H/o HT, steroid intake, oral contraceptive pills, trauma, tumor, treatment were carefully sought.

Ophthalmic evaluation:

Visual acuity, tension, colour vision, field, fundus examination with direct ophthalmoscope, indirect ophthalmoscope and +90 D lens were noted. A detailed neurological examination was done in each patient which included examination of higher functions, cranial nerves, sensory system, motor reflex and cerebellum.

Investigations like blood Hb%, TC, DC, blood sugar, CT brain, MRI brain were done in all patients.

OBSERVATION

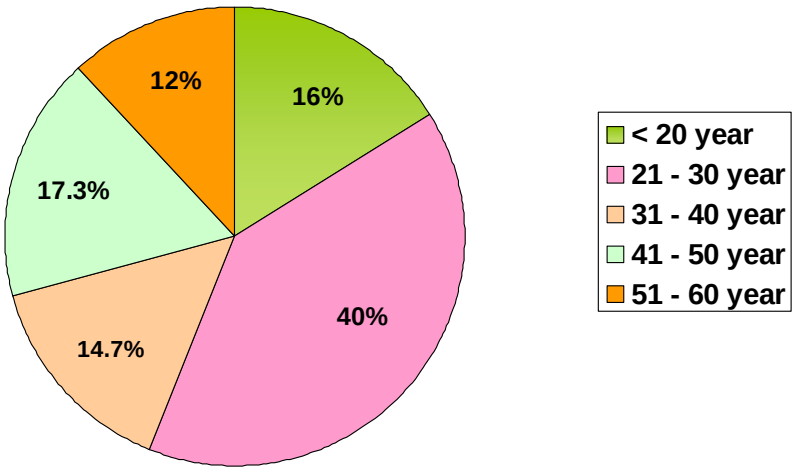
1. AGE:

The 75 patients studied were classified according to age and the percentage of papilloedema in each age group was studied

Table 3

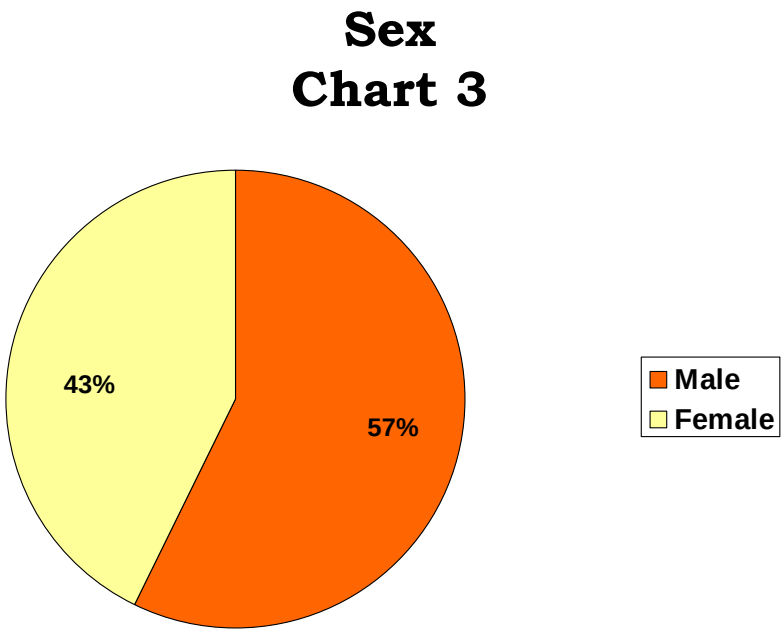
Age group	No of cases	Percentage
Less than 20 years	12	16%
21 – 30 years	30	40%
31 – 40 years	11	14.7 %
41 – 50 years	13	17.3%
51 – 60 years	9	12%

Age group
Chart 2



2. SEX:

Out of the 75 patients studied 43 of them were females (57.3%) and 32 of them were males (42.8%)

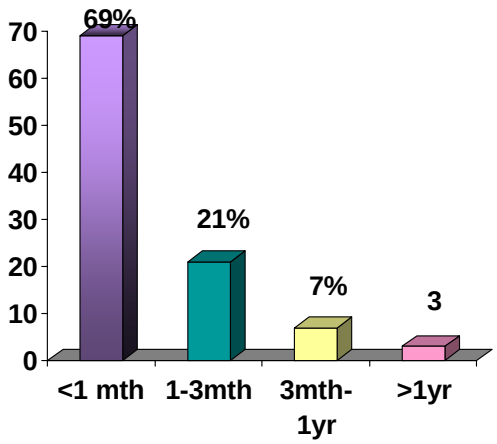


3. DURATION OF ILLNESS:

Table 4

Duration of illness	No of cases	Percentage
Acute (< 1month)	52	69%
Sub acute(1-3month)	16	21%
Chronic (3 month-1year)	5	7%
Very chronic (> 1 year)	2	3%

Duration of illness
Chart 4

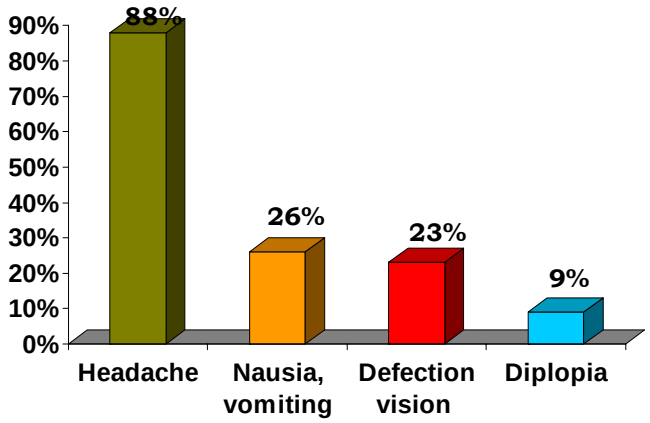


4. PRESENTING SYMPTOMS:

Table 5

Symptoms	No of cases	Percentage
Head ache	66	88%
Nausea, Vomiting	33	26%
Defection vision	29	23%
Diplopia	11	9%

Presenting symptoms
Chart 5

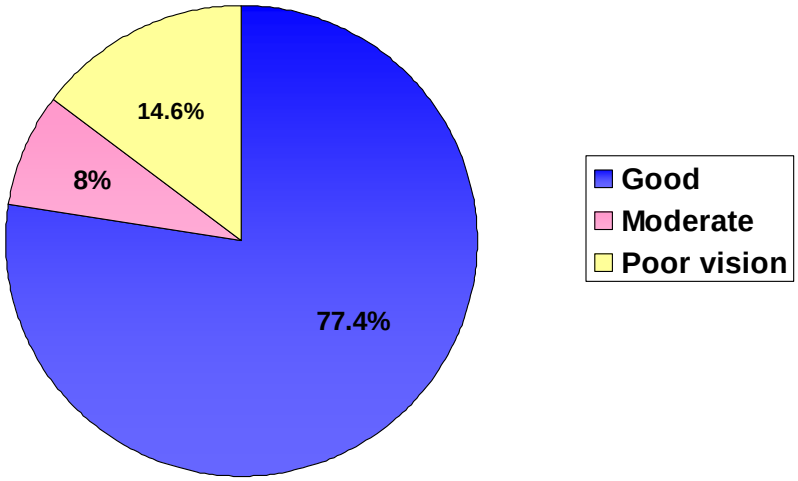


5. VISUAL LOSS:

Table 6

Visual loss	No of cases	Percentage
Good (>6/18)	58	77.4%
Moderate(6/60 – 6/18)	6	8%
Poor vision (<6/60)	11	14.6%

Visual loss
Chart 6

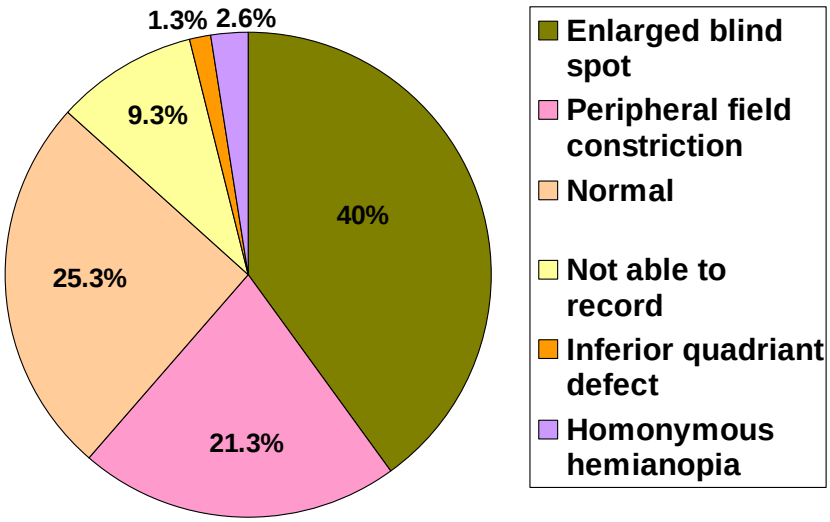


6. VISUAL FIELD DEFECT:

Table 7

Visual field defect	No of cases	Percentage
Enlarged blind spot	30	40%
Peripheral field constriction	16	21.3%
Normal	19	25.3%
Not able to record	7	9.3%
Inferior quadrant defect	1	1.3%
Homonymous hemianopia	2	2.6%

Visual field defect
Chart 7



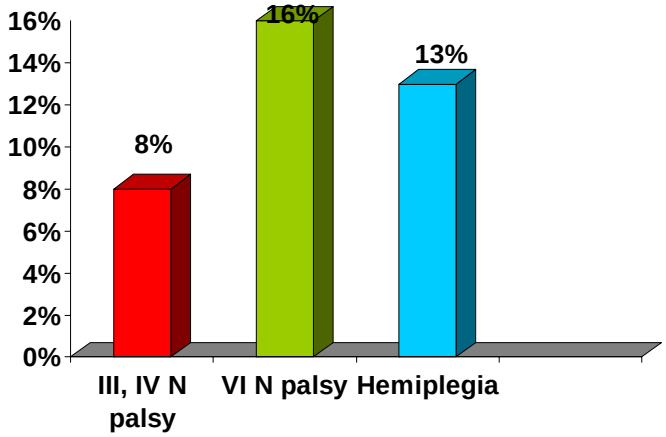
7. RAPD noted in 6 cases

8. LOCALISING NEUROLOGICAL SIGNS:

Table 8

Localising neurological signs	Percentage
III, IV N palsy (including Webers syndrome)	8%
VI N palsy	16%
Hemiplegia	13%

Localising Neurological signs
Chart 8

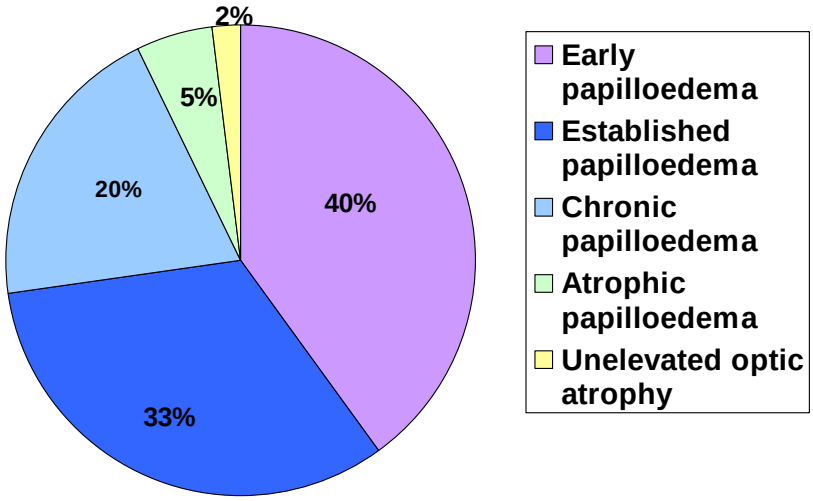


9. OPHTHALMOSCOPIC EXAMINATION:

Table 9

Ophthalmoscopic examination	No of cases	Percentage
Early papilloedema	30	40%
Established papilloedema	25	33%
Chronic papilloedema	15	20%
Atrophic papilloedema	3	5%
Unilateral optic atrophy with contra lateral papilloedema (Foster Kennedy syndrome)	2	2%

Ophthalmoscopic examination
Chart 9

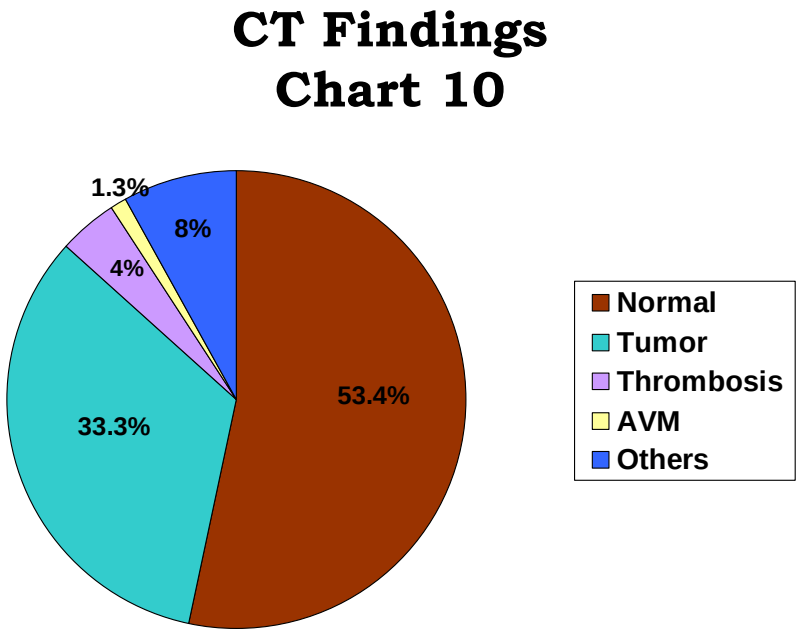


10. CT FINDINGS:

Table 10

CT findings	No of cases	Percentage
Normal	40	53.4%
Tumour	25	33.3%
Thrombosis	3	4%
AVM	1	1.3%
Others	6	8%

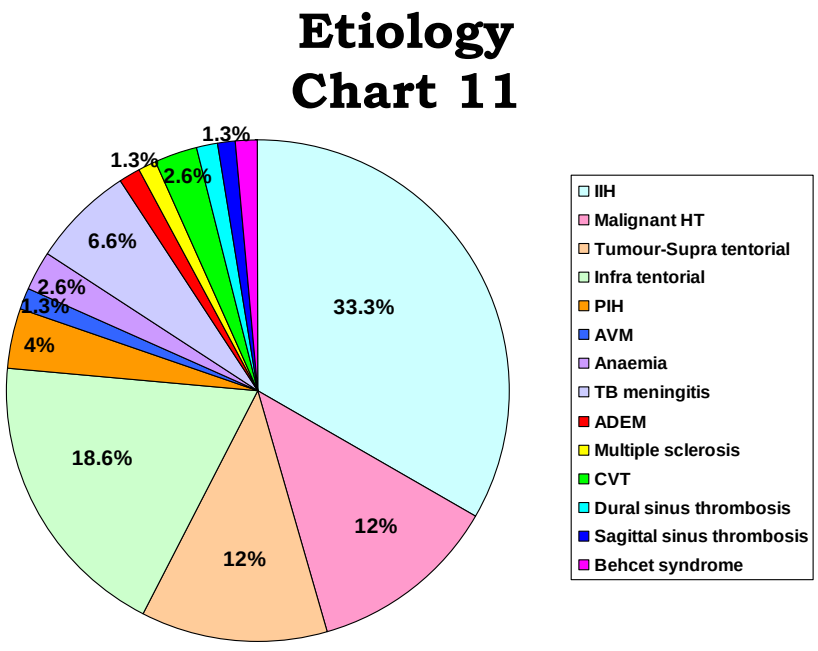
CT was normal in 40 patients of which 25 had pseudo tumour cerebri, 9 had malignant hypertension, 3 had PIH and 2 had iron deficiency anaemia.



11. ETIOLOGY:

Table 11

Etiology	No of cases	Percentage
Idiopathic intra cranial hypertension	25	33.3%
Malignant hypertension	9	12%
Tumour-supratentorial	9	12%
Infratentorial	14	18.6%
PIH	3	4%
AVM	1	1.3%
Anaemia	2	2.6%
TB meningitis	5	6.6%
ADEM	1	1.3%
Multiple sclerosis	1	1.3%
CVT	2	2.6%
Dural sinus thrombosis	1	1.3%
Sagittal sinus thrombosis	1	1.3%
Behcet syndrome	1	1.3%



DISCUSSION

1. Age:

Out of the 75 cases 16% occurred in the first 2 decades, 40% in the 3rd decade, 14.7% in 4th decade and 12% beyond 5th decade of life.

These results are similar to the study by Subramaniam et. al. who reported 52% in the 3rd decade[11].

2. Sex:

57.3% were females and 42.8% were males. In the study of Subramaniam et al, they found 55% of males and 42% females.

3. Duration of illness:

Similar to the other study mentioned above, 69% were acute, 21% sub acute, 7% chronic and 3% very chronic illness.

4. Presenting symptoms:

Head ache was the commonest symptom in 88% of cases, similar to 85% in the study of Subramanian, et al[11] Nausea and vomiting was present in 26% as compared to 56% in Subramaniam et al study. This can probably be explained by Benign Intra Cranial Hypertension being our predominant etiology (40%) as compared to SOL 73% in their study.

5. Visual loss:

Fall of vision from 6/18 – 6/60 was noted in 8% and <6/60 in 14.6% similar to those in other studies. The 11 patients with poor vision had etiologies of dural venous thrombosis, cortical vein thrombosis, recurrent CP angle tumour, Neuro cysticercosis, fronto parietal perisylvian anaplastic astrocytoma grade 3, supra tentorial space occupying lesion and sphenoid ridge meningioma.

Diplopia was observed in 24% of patients of which 16% were due to V1nerve palsy (false localizing sign) and 8% were due to III & IV nerve palsy[1].

6. Visual field defect:

Enlarged blind spot noted in 42.6% of cases, peripheral field constriction 21.3%, no field defect was noted in 18.6% of cases (13) Field could not be tested in 14.6% because of poor vision.

Hemianopia was observed in 2 cases which had associated signs (i) cerebellar signs (Neuro cysticercosis – multiple cyst in cerebellum and cerebral cortex) (ii) Weakness of right upper and lower limb (mass in parietal lobe).

7. Relative afferent pupillary defect: (RAPD)

RAPD was noted in 4 cases, of which secondary optic atrophy occurred in 3 cases. One case with BIH came to OPD with V/A, RE 6/60, LE –CFCF, RAPD was present in the left eye, LP was done – no cells in CSF (4ml drained to decrease ICT) she was treated with Inj.Solumedrol and lasix for 3 days. After 1 week papilloedema resolved, her vision improved to 6/18 and after 1 week to 6/6 both eyes.

Absolute afferent pupillary defect in both eyes was noted in 2 cases of Dural venous thrombosis.

8. Neurological signs of localizing value were detected in 37% of cases as compared to 70% in Subramaniam et al study.

IV N palsy was noted in 16% cases and other cranial nerve palsy were noted in 8% Hemiplegia in 13% (14)

14.6% of patients who presented at OPD directly were detected as papilloedema and referred to neurosurgery department for diagnosis of SOL 2 cases were diagnosed as having granuloma parietal lobe, and 1 having tempero parietal SOL, 7 cases were diagnosed as BIH.

9. Ophthalmoscopic examination:

40% of the 75 patients showed early papilloedema, 34.7% showed established papilloedema, 20% showed chronic papilloedema, 5.3% atrophic papilloedema. 2 cases of Foster

Kennedy syndrome were also detected with one eye papilloedema and other eye optic atrophy, etiology being parietal SOL and sphenoid ridge meningioma.

This was similar to Subramanian et al study[11].

10. CT findings:

CT was normal in 53.4% of which 25 patients had Idiopathic intra cranial hypertension (33%), 9 had malignant hypertension (12%), 3 had PIH (4%), 2 had iron deficiency anaemia (2.6%), SOL constitute 33.3%

4 out of the 25 Idiopathic intra cranial hypertension patients had no complaints and papilloedema was detected on routine ophthalmic examination (15)

11. Etiology:

Idiopathic intra cranial hypertension was found to be the commonest cause of papilloedema in my study. 13 patients with Idiopathic intra cranial hypertension were medically managed and showed drastic improvement in symptoms.

2 patients underwent lumbar puncture to reduce the CSF pressure. One was of unknown cause presenting with severe visual loss, the other was a steroid induced BIH, presenting with severe head ache. Treatment relieved their visual loss and head ache.

Tumours:

Out of the 23 patients with intra cranial tumour 32% were supra tentorial and 50% were infra tentorial tumours which is in concurrence with study (11) by Subramaniam et al.

Gliomas constituted 9.3% of the papilloedema patients, meningioma 6.6%, Neurocysticercosis 2.6%, CP angle tumour 2.6% clival chordoma 1.3% (17)

5 cases were treated surgically by craniotomy and decompression, 2 cases treated with both surgery and radiotherapy. 6 cases died prior to surgery 2 cases referred to higher centre for further management. 10 cases were lost to follow up.

Anaemia:

2 cases of anaemia manifested with papilloedema. Papilloedema in iron deficiency anaemia is mainly due to cerebral ischaemia and cerebral edema and it is very rare. This is in concurrence with study by V.Biocusse, N.Newman et al (19).

TB meningitis:

TB meningitis occurred in 6.6% of cases. They were treated with ATT and all showed improvement after treatment. This low incidence is as compared to Subramanian et al study can be explained by the MDT (Multi Drug Therapy) and early diagnosis by CT / MRI / Anti TB IgM availed in the recent times.

Malignant HT:

12% of the papilloedema patient had malignant hypertension, similar to Shelbure et al study[12].

Behcet syndrome:

One of the patient in my study was a case of Behcet syndrome with DVT on treatment which is in concurrence with a case report by Panir NM, Kansu T et al[23] .

Cortical Vein Thrombosis:

4 patients with Cortical Vein Thrombosis in my study presented with sudden, visual loss in both eye and multiple cranial nerve palsies[26] .

CONCLUSION

1. Papilloedema is most common in the 3rd decade of life and rare after 50 years of age. There is a female preponderance.
2. Headache is the commonest symptom and vomiting is rare according to my study.
3. Poor vision at presentation and late detection of papilloedema are factors responsible for poor visual outcome.
4. Symptoms of vomiting and diplopia are infrequent but important indicators of papilloedema.
5. Enlarged blind spot is the commonest field defect.
6. Idiopathic intra cranial hypertension was the commonest etiology for papilloedema in my study.
7. Infra tentorial tumours are the commonest intra cranial space occupying lesions causing papilloedema.
8. All patients visiting the ophthalmology department must have a fundus examination to detect papilloedema especially those presenting with headache.
9. Behcet syndrome and iron deficiency anaemia have to be born in mind as rare but proven causes of papilloedema.

RECOMMENDATION & PREVENTIVE PROSPECTS

1. All the patients with headache should be evaluated ophthalmoscopically to look for papilloedema.

2. Early diagnosis and prompt treatment may relieve symptoms.

3. In chronic cases papilloedema resolves into optic atrophy and visual loss is the outcome. So early treatment is necessary to retain vision.

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PROFORMA

Name : Age / Sex :

IP / OP No : Occupation:

Address : Contact No:

1) PRESENTING COMPLAINTS:

Headache / Nausea / Vomiting

Visual Complaints :

Diplopia / Transient Blurring of vision / photopsia / gradual loss of vision

Neurological Complaints:

Loss of consciousness / motor weakness / sphincter control / personality disturbance / Head Trauma / Seizures.

ENT Complaints :

Speech / hearing problem / Nasal regurgitation / CSF rhinorrhea / Bleeding per ear / Tinnitus.

2) DURATION OF ILLNESS

3) PAST HISTORY

DM / HT / IHD / BA / TB / Thyroid Ds / Arthritis / Similar episodes.

4) PERSONAL HISTORY

Smoker / alcoholic / Veg / Non-veg / Exposure to pet animals

Exposure to STD / Abortion – In female patients – Obstetric history – menstrual cycle, PIH.

5) FAMILY HISTORY

6) TREATMENT / DRUG HISTORY :

Vit A / OCP / Nalidixic acid, steroids / Medical or Surgical / ENT/Neck surgery.

7) GENERAL EXAMINATION

Conscious / Orientation / Nourishment / Anaemia Jaundice / clubbing / Generalised Lymphadenopathy / Pedal edema / Neurocutaneous markers / Congenital anomalies.

8) SYSTEMIC EXAMINATION

CVS - BP - PR

RS-

P/A -

CNS -

OPHTHALMIC EXAMINATION

Head posture

Facial symmetry

Exophthalmos / Enophthalmos R.E L.E

Eye position

Eye lids

Ocular movements

Conjunctiva

Cornea

Anterior Chamber

Iris

Pupil

- Size
- Shape
- Light reflex
  - Direct
  - Consensual
- Near reflex

Lens

Anterior Vitreous phase Posterior vitreous

FUNDUS Both Direct & Indirect

- Media R.E L.E
- Disc
  - Size
  - Shape
  - Margins
  - Colour
  - Cup: Disc ratio

Lamina cribrosa

Vessels on the disc

Peripapillary region

Edema

Folds

Laminar separation

Hemorrhages

Others

Vessels

Sheathing

Pulsations

A: V ratio

Periphery

Hemorrhages

Exudates

Others

Macula

Fundus Diagnosis:

Visual Acuity

Distant

Without correction

With correction

Nearvision

Colour vision

Retinoscopy

Intraocular Tension

Gonioscopy

Visual Fields

Central Peripheral

Diplopia Charting

Hess charting

NEUROLOGICAL EXAMINATION

Higher functions

Cranial nerves R.E L.E

Motor system

Sensory system

Cerebellar system

Reflexes

Cardiovascular system Respiratory system

Gastrointestinal system

E.N.T. Examination: Endocrine system

PROVISIONAL DIAGNOSIS

INVESTIGATIONS

Hematology:

Hb% TC DC

ESR

RBC count Platelet count

Mantoux: Blood VDRL ELISA

Blood sugar: Fasting Post – prandial

C.S.F analysis (if any)

Urine – Albumin Sugar Motion-Ova Cyst

Radiological examination:

X-Ray Skull, Orbit, Chest,

CT Scan

M.R.I. Scan

Ultra sound Examination

Fundus Fluorescin Angiography

FINAL DIAGNOSIS

Treatment

Medical

Surgical

FOLLOW UP.

MASTER CHART

S.No	Age	Sex	Head ache	Nausea & Vomiting	Duration	Past H/o TB BA	RAPD	Cranial nerve involvement					V/A	Fundus	CT	Field	Etiology
								III	IV	V	VI	Hemi plegia					
1	26	F	Y	Y	3 days	Y	N	N	N	N	N	N	6/6	Chronic	N	EBS	IIH
2	42	M	Y	N	2 weeks	N	N	N	N	N	N	N	6/6	Early	Mass	EBS	SOL
3	55	F	Y	Y	10 days	N	N	N	N	N	N	N	6/6	Early	Mass	EBS	SOL
4	45	M	Y	N	3 months	N	N	N	N	N	N	N	6/6	Established	Mass	N	SOL
5	5	M	Y	Y	3 months	N	N	N	N	N	N	N	6/6	Early	Mass	EBS	SOL
6	13	F	Y	Y	15 days	N	N	N	N	N	N	N	6/18	Early	Mass	EBS	SOL
7	30	F	Y	Y	15 days	N	N	N	N	N	N	N	6/6	Established	Mass	EBS	SOL
8	25	F	Y	N	2 weeks	N	Y	N	N	N	Y	Y	6/6	Established	N	EBS	TBM
9	30	M	Y	N	1 week	N	N	N	N	N	N	N	6/6	Early	N	EBS	IIH
10	15	F	Y	Y	3 months	N	N	Y	Y	Y	Y	N	6/24	Established	Mass	EBS	SOL
11	40	M	N	N	20 days	N	N	N	N	N	N	N	6/60	Early	Mass	EBS	SOL
12	20	F	Y	N	1 week	N	N	N	N	N	N	N	6/6	Early	N	EBS	ADEM
13	29	F	Y	Y	15 days	N	N	N	N	N	N	N	6/6	Chronic	N	EBS	Anaemia
14	16	M	N	Y	2 weeks	N	N	N	N	N	N	N	6/6	Early	N	EBS	IIH
15	23	F	Y	Y	1 month	N	N	N	N	N	N	N	6/6	Early	N	EBS	PIH

MASTER CHART

S.No	Age	Sex	Head ache	Nausea & Vomiting	Duration	Past H/o TB BA	RAPD	Cranial nerve involvement					V/A	Fundus	CT	Field	Etiology
								III	IV	V	VI	Hemi plegia					
16	34	F	Y	Y	6 months	N	N	N	N	N	N	N	6/6	Established	N	EBS	IIH
17	54	F	Y	Y	1 week	N	N	N	N	N	N	N	6/36	Early	Mass	EBS	SOL
18	24	F	Y	Y	1 week	N	N	N	N	N	N	N	6/6	Early	N	EBS	IIH
19	13	F	Y	N	1 week	N	N	N	N	N	N	N	6/6	Early	N	PFC	IIH
20	24	F	Y	Y	2 week	N	N	N	N	N	N	N	6/6	Early	N	PFC	PIH
21	57	M	Y	N	2 months	N	N	Y	Y	N	N	Y	6/6	Established	N	EBS	IIH
22	45	F	Y	N	6 months	N	N	N	N	N	N	N	6/6	Early	N	PFC	IIH
23	32	F	Y	Y	1 week	N	N	N	N	N	N	N	6/6	Early	N	N	IIH
24	39	F	Y	N	6 days	N	N	N	N	N	N	N	6/6	Early	Mass	PFC	SOL
25	13	F	Y	Y	1 month	N	N	N	N	N	N	N	6/6	Eearly	Mass	PFC	SOL
26	50	F	Y	Y	4 months	N	N	N	N	N	N	N	6/6	Established	N	PFC	IIH
27	30	F	Y	N	6 months	N	N	N	N	N	N	N	6/6	Chronic	N	PFC	IIH
28	50	M	N	N	6 months	N	N	Y	Y	Y	N	Y	CFCF	Atrophic	DLV	NAT	SOL
29	28	F	Y	Y	4 months	N	N	N	N	N	N	N	6/6	Early	N	EBS	IIH
30	20	F	Y	Y	10 days	N	N	N	N	N	N	N	6/6	Established	N	N	IIH

MASTER CHART

S.No	Age	Sex	Head ache	Nausea & Vomiting	Duration	Past H/o TB BA	RAPD	Cranial nerve involvement					V/A	Fundus	CT	Field	Etiology
								III	IV	V	VI	Hemi Plegia					
31	30	F	Y	N	1 month	Y	N	N	N	N	Y	Y	6/6	Chronic	Mass	EBS	TBM
32	26	M	N	N	4 years	N	N	N	N	N	N	N	1/60	Atrophic	Mass	NAT	SOL
33	14	F	Y	N	2 weeks	N	N	N	N	N	Y	Y	6/6	Established	N	PFC	TBM
34	25	F	Y	N	2 weeks	N	N	N	N	N	N	N	6/6	Established	N	N	IIH
35	5	M	Y	N	2 months	N	N	N	N	N	N	N	6/6	Established	N	N	IIH
36	28	F	Y	N	2 weeks	N	N	N	N	N	N	N	6/6	Established	AVM	N	AVM
37	22	M	Y	N	10 days	N	N	N	N	N	N	N	6/6	Early	N	EBS	HT
38	32	F	Y	N	1 year	N	N	N	N	N	N	N	6/60	Chronic	Mass	PFC	SOL
39	43	M	Y	N	3 days	N	N	N	N	N	N	N	6/6	Early	N	N	IIH
40	24	M	Y	N	2 days	N	N	N	N	N	N	N	6/6	Eearly	N	PFC	HT
41	55	M	Y	N	3 month	N	N	N	N	N	Y	Y	6/36	Established	N	N	IIH
42	43	M	N	N	1 week	N	N	N	N	N	N	N	6/6	Early	N	PFC	HT
43	27	F	N	N	1 week	N	N	N	N	N	N	N	6/6	Early	N	PFC	PIH
44	9	F	Y	N	10 days	N	N	N	N	N	N	N	6/6	Established	N	PFC	HT
45	29	M	Y	Y	1 week	N	N	N	N	N	N	N	6/6	Established	N	EBS	CVT

MASTER CHART

S.No	Age	Sex	Head ache	Nausea & Vomiting	Duration	Past H/o TB BA	RAPD	Cranial nerve involvement					V/A	Fundus	CT	Field	Etiology
								III	IV	V	VI	Hemi plegia					
46	22	M	Y	N	2 weeks	N	N	N	N	N	N	N	6/6	Established	N	N	Behcet
47	58	M	Y	N	2 weeks	N	N	N	N	N	N	N	6/6	Established	N	EBS	HT
48	41	M	Y	N	6 days	N	N	N	N	N	N	N	6/6	Early	N	PFC	IIH
49	40	F	Y	N	1 month	N	Y	N	N	N	Y	Y	1/60	Atrophic	Mass	NAT	SOL
50	42	M	N	N	1 week	N	N	N	N	N	N	N	CFCF	Established	N	Others	IIH
51	45	F	Y	Y	2 weeks	N	N	N	N	N	N	N	6/6	Established	N	EBS	HT
52	25	F	Y	N	15 days	N	N	N	N	N	N	N	6/6	Established	N	N	Anaemia
53	13	F	Y	N	1 week	N	N	N	N	N	Y	N	6/6	Early	N	N	IIH
54	24	F	Y	Y	2 weeks	N	N	N	N	N	N	N	6/6	Early	N	N	HT
55	50	M	Y	N	3 months	N	N	N	N	N	N	N	1/60	Atrophic	DLV	NAT	SOL
56	47	F	Y	N	3 months	Y	N	Y	Y	N	Y	N	6/60	Established	Mass	PFC	SOL
57	27	F	Y	N	6 months	N	N	N	N	N	N	N	HM	Chronic	Mass	NAT	SOL
58	54	M	Y	Y	45 days	N	N	N	N	N	Y	Y	6/6	Early	N	N	TBM
59	50	F	Y	Y	6 month	N	N	N	N	N	N	N	6/6	Chronic	N	N	IIH
60	30	M	N	N	2 months	N	N	N	N	N	N	N	1/60	Early	Mass	NAT	SOL

MASTER CHART

S.No	Age	Sex	Head ache	Nausea & Vomiting	Duration	Past H/o TB BA	RAPD	Cranial nerve involvement					V/A	Fundus	CT	Field	Etiology
								III	IV	V	VI	Hemi Plegia					
61	40	M	Y	Y	25 days	Y	N	N	N	N	N	N	6/6	Chronic	SOL	EBS	SOL
62	28	F	Y	Y	25 days	N	Fixed	Y	Y	N	Y	Y	4/60	Chronic	DVT	NAT	DVT
63	50	F	Y	Y	2 years	N	N	N	N	N	Y	N	6/6	Chronic	SOL	N	SOL
64	84	M	Y	Y	45 days	N	N	N	N	N	N	N	6/6	Established	SOL	N	HT
65	30	M	Y	Y	60 days	HIV +ve	N	N	N	N	N	N	1/60	Chronic	Mass	NAT	SOL
66	29	M	Y	Y	1 week	N	N	N	N	N	Y	Y	6/6	Established	N	EBS	TBM
67	38	M	Y	Y	1 week	HIV +ve	N	N	N	N	N	N	6/6	Established	N	EBS	IIH
68	60	M	Y	Y	10 days	N	N	N	N	N	N	N	6/6	Established	SDH	EBS	IIH
69	25	F	Y	Y	1 week	N	N	N	N	N	Y	Y	6/6	Early	CVT	EBS	DVT
70	28	F	Y	Y	20 days	N	Y	Y	Y	N	Y	N	HM	Chronic	DVT	NAT	DVT
71	24	M	Y	N	1 week	N	N	N	N	N	N	N	6/6	Early	N	N	IIH
72	40	F	Y	N	2 days	N	N	N	N	N	N	N	6/6	Chronic	SOL	N	SOL
73	35	M	Y	N	2 months	N	N	N	N	N	N	N	3/60	Chronic	SOL	N	SOL
74	40	M	Y	N	25 days	N	N	N	N	N	N	N	6/6	Established	N	EBS	IIH
75	50	F	Y	Y	1 month	N	N	N	N	N	N	N	6/6	Chronic	SOL	PFC	SOL

HM – Hand movement
CFCF – Counting finger close to face
IIH – Idiopathic Intra cranial Hypertension
SOL – Space Occupying lesion
TBM – Tuberculous Meningitis

Early - Early papilloedema
Established - Established papilloedema
Chronic - Chronic papilloedema
Atrophic - Atrophic papilloedema
DVT - Dural Venous Thrombosis

EBS – Enlarged Blind Spot
NAT – Not Able to Test
SDH – Sub Dural Haemorrhage